Pol J Pathol 2011; 1: 50–59

Tumours and tumour-like lesions of the spinal canal and spine. A review of 185 consecutive cases with more detailed close-up on some chosen pathologies

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Objectives: Spinal canal tumours constitute a minor part of CNS invading neoplasm. However, due to their damaging influence on the spinal cord and the spinal roots, they cause serious clinical problems and can lead to severe disability.
The aim of this study is to review material collected on tumours of the spine and the spinal canal at the Department of Neuropathology over the past ten years.
Material and methods: One hundred and eighty five histopathological examinations of spinal tumours were evaluated between August 1997 and August 2007. The group of patients included 94 females and 91 males between the age of 18 and 79 years with a mean age of 53.
Results: Apart from typical intraspinal tumours (i.e. astrocytomas and ependymomas), and extraspinal tumours, (i.e. meningiomas, schwannomas, neurofibromas), rare neoplastic and non-neoplastic tumour-like changes occur in the same localizations. These rare conditions include: capillary haemangioma, paraganglioma of filum terminale, meningeal gliomatosis, different variants of cysts such as the dermoid cyst, synovial cyst and aneurysmatic bone cyst, neoplastic and non-neoplastic bone tumours like the giant cell tumour, chordomas, and intramedullary metastatic carcinomas.
Conclusions: This paper presents and discusses spinal lesions from collected data with special attention paid to the rare conditions, which are reviewed in more detail.

Key words: spinal canal, pathology, capillary haemangioma, paraganglioma, meningeal gliomatosis, dermoid cyst, synovial cyst, intramedullary metastatic carcinoma.

Introduction

Spinal canal and spine tumours constitute a heterogeneous group of pathologies. Some of them, like intramedullary tumours and tumours of the meninges and spinal roots, are classified as tumours of the Central Nervous System according to the newest WHO classification [1]. Tumours of other structures of the spine, like bone tumours, chordomas, and many types of cysts are not included in this classification. However, in the everyday practice of a neurosurgeon or a neuropathologist, removing and diagnosing a tumour localized anywhere in the vertebral column should comprise all types of tumours and tumour-like conditions. This problem has been addressed by Van Goethem, whose classification includes other types of tumours (Table I) [2].

An important anatomical classification, especially from a neurosurgical point of view, differentiates extradural and intradural tumours. Intradural tumours contain intra- and extraspinal tumours. Another important and practical differentiation separates primary and

Table I. WHO Classification of spinal tumours in modification of Van Goethem *et al.*

NEUROEPITHELIAL TUMOURS		
	Astrocytic tumours (glial tumours)	Fibrillary astrocytomas (WHO grade II) Anaplastic (malignant) astrocytoma (WHO grade III) Glioblastoma multiforme (WHO grade IV) Pilocytic astrocytoma (WHO grade I) Pleomorphic xanthoastrocytoma (WHO grade II)
	Oligodendroglial tumours	Oligodendroglioma (WHO grade II) Anaplastic oligodendroglioma (WHO grade III)
	Ependymal cell tumours	Ependymoma (WHO grade II) Anaplastic ependymoma (WHO grade III) Myxopapillary ependymoma (WHO grade I) Subependymoma (WHO grade I)
	Mixed gliomas	Mixed oligoastrocytoma (WHO grade II) Anaplastic oligoastrocytoma (WHO grade III)
	Neuronal and mixed neuronal -glial tumours	Gangliocytoma (WHO grade I) Ganglioglioma (WHO grade I/II) Anaplastic (malignant) ganglioglioma (WHO grade III) Desmoplastic infantile ganglioglioma (WHO grade I) Dysembryoplastic neuroepithelial tumour (DNET) (WHO grade I) Paraganglioma (WHO grade I)
	Neuroblastic	Neuroblastoma
	Embryonal tumours	Ependymoblastoma (WHO grade IV)
Peripheral nerve tumours		
	Schwannoma (neurinoma, neurilemmoma) (WHO grade I) Neurofibroma (WHO grade I) Malignant peripheral nerve sheath tumour (WHO grade III/IV)	
Hematopoietic tumours		
	Primary malignant lymphomas Plasmacytoma Leukaemia	
Germ cell tumours		
	Embryonal carcinoma Teratoma Mixed germ cell tumours	
Tumours of the meninges		
	Meningothelial tumours	Meningioma (WHO grade I) Atypical meningioma (WHO grade II) Anaplastic meningioma (WHO grade III)
	Mesenchymal, non-meningothelial tumours	Lipoma Angiolipoma Hibernoma Fibrosarcoma Malignant fibrous histiocytoma Chondroma, chondrosarcoma Osteoma, osteosarcoma Osteochondroma Haemangioma Hemangiopericytoma
	Melanocytic lesions	Melanocytoma **Malignant melanoma** Meningeal melanocytosis
	Tumours of unclear origin	Hemangioblastoma

Table I. cont.

Metastatic tumours	
Primary bone tumours	
	Aneurysmal bone cyst
	Chordoma
	Chondrosarcoma
	Ewing sarcoma
	Fibrosarcoma
	Giant cell tumour
	Haemangioma
	Histiocytosis
	Lymphoma
	Myeloma
	Osteoid osteoma
	Osteoblastoma
	Osteosarcoma

Table II. The histopathological diagnoses of cases operated as tumours of the spinal canal and spine in the material of the Department of Neuropathology, Jagiellonian University Medical College

TUMOUR	NUMBER OF CASES	%
Meningioma	65	35.13
Schwannoma	31	16.75
Metastatic tumours	24	12.97
Ependymoma	19	10.27
Malignant lymphoma	12	6.48
Astrocytoma	8	4.32
Angioma	5	2.7
Neurofibroma	4	2.16
Plasmacytoma	2	2.16
PNET	2	2.16
Tumours of the hematopoietic system	2	2.16
Chordoma	1	0.54
Dermoid cyst	1	0.54
Ewing sarcoma	1	0.54
Extraparenchymal spinal neuroglial cyst	1	0.54
Giant cell tumour of bone	1	0.54
Lipoma	1	0.54
Malignant fibrous histiocytoma	1	0.54
Meningeal gliomatosis	1	0.54
Non specific inflammatory changes	1	0.54
Paraganglioma	1	0.54
Synovial cyst	1	0.54
Total	185	100

metastatic tumours. Statistics on all spinal canal tumours from different reports show extremely different frequencies of occurrence in particular anatomical compartments as follows: extradural tumours of 20-78%, intradural extraspinal tumours of 18-60%, and intraspinal tumours of 4-20%. Among the primary CSN tumours, spinal cord tumours account for approximately 15%. In the case of astrocytoma, the ratio of the intracranial tumours to spinal canal tumours is 10 : 1 and for ependymomas between 3-20 : 1 [3-7].

The anatomy of the spinal canal is especially important in understanding the pathology of tumours at this localization. Anatomical conditions strictly determine the pathophysiology of a spinal canal tumour. The stiff bone structure of the spinal canal and the close apposition of nervous and bone elements causes an additional mass, like a tumour, to produce an increased intracanal pressure. This in turn damages the nervous structures by direct pressure or compression of blood vessels and leads to circulatory disturbances in the spinal cord and roots. In the first line, veins are compressed, which results in venous stasis and oedema. Higher pressure may obliterate the terminal arterial branches and form ischemic foci, often remote from the place of compression [8].

The symptomatology of spinal canal tumours depends on their localization and histology. The symptoms usually develop gradually and insidiously. Initially, pain symptoms and sensory disturbances are dominant. Subsequently, motor deficits appear. At this point it has to be noted that the spinal cord has great compensation abilities, enabling, especially tumours with slow growth, attainment of a relatively large size mass with minor or even without symptoms [8, 9].

The aim of this study is to present material collected about spinal canal and spine tumours in the Department of Neuropathology at the Jagiellonian University Medical College. Furthermore, this paper

Table III. Localization of the spinal canal and spine tumours with regard to three particular compartments (The order of tumours is the same as in Table II)

TUMOUR	INTRAMEDULLAR	INTRADURAL EXTRAMEDULLAR	EXTRADURAL
Meningioma	0	61	4
Schwannoma	0	28	7
Metastatic tumours	1	0	23
Ependymoma	9	10	0
Malignant lymphoma	0	0	12
Astrocytoma	8	0	0
Angioma	4	1	0
Neurofibroma	0	4	1
Plasmacytoma	0	0	2
PNET	2	0	0
Tumours of the hematopoietic system	0	0	2
Synovial cyst	0	0	1
Extraparenchymal spinal neuroglial cyst	0	1	0
Paraganglioma	0	1	0
Meningeal gliomatosis	0	1	0
Dermoid cyst	0	1	0
Giant cell tumour of bone	0	0	1
Malignant fibrous histiocytoma	0	0	1
Ewing sarcoma	0	0	1
Lipoma	1	0	0
Chordoma	0	0	1
Non specific inflammatory changes	1	0	0
Total*	26 (14.05%)	108 (58.37%)	56 (36.75%)

**The sum of the pathologies in the particular compartments exceeds total number of cases (185) because some tumours were localized in more than one compartment (intradurally and extradurally)*

discusses in a more detailed fashion selected rare pathological lesions of the spinal canal.

Material and methods

We included into the analysis 185 consecutive histopathological examinations of spine tumours and non-neoplastic lesions made during a ten years' period from August 1997 to August 2007 in the Department of Neuropathology at the Jagiellonian University Medical College. There were 94 females and 91 males between the age of 18 and 79 years (mean age 53 years) in this study.

Results

The most common tumours found in the examination of data were meningiomas. They constituted 32.5% of the entire group of two hundred tumours. The spectrum of different meningioma subtypes is presented in Table IV. The second most common type of tumours from the group was schwannoma, which constituted 15.5%. The third largest group (11.5%) was formed by metastatic carcinomas. However, if counted together, tumours of glial origin (19 ependymomas, 8 astrocytomas and 1 meningeal gliomatosis) would occupy the third place when considering frequency (28 cases in all, 14% of the whole set). Investigation of gathered material revealed some rare pathologies. Spinal canal tumours such as: capillary haemangioma, paraganglioma, meningeal gliomatosis, dermoid cyst, synovial cyst, and intramedullary metastatic carcinoma belong to this category. These tumours are presented below in a more detailed fashion. Some typical bone tumours were also diagnosed including giant

Table IV. Subtypes of the tumours

MENINGIOMA	NUMBER OF CASES
Meningothelial	31
Psammomatous	18
Transitional	7
Angiomatous	3
Clear cell	2
Fibrous	2
Atypical	1
Malignant	1
Ependymoma	
WHO II	15
WHO I – myxopapillary	4
Astrocytoma	
Fibrillary	6
Pilocytic	2

cell tumour of bone, malignant fibrous histiocytoma, Ewing's sarcoma, and chordoma. The number and percentage of all pathologies are listed in Table II. The most common segment invaded by the tumour was the thoracic region (Table VIII). The different localization about the dura mater for each type of the tumour is shown in Table III. Some schwannomas (4 cases in our material) and neurofibromas grow intra- and extradurally. Metastatic carcinomas most likely occur extradurally (22 cases). One metastatic tumour was placed in the intramedullar region (see case presentation 6 below). Our primary focus regarding the spread of carcinomas is presented in Table V. Ependymomas were found intramedullary in 9 cases and extramedullary-intradurally in 10 cases. Four cases of myxopapillary ependymomas were localized intradurally around the cauda equina (Table IV). Only eight astrocytomas were found (Table IV). The case of meningeal gliomatosis was accounted for and presented separately. Moreover, some physiological structures were found like nucleus pulposus, spinal root, ligament tissue, and fibrous tissue, these are not included in the statistics.

Our report does not include pathologies like nucleus pulposus, subdural hematoma, nonspecific inflammatory changes, or just samples of histologically apparently normal tissue like: fragments of the spinal root, nondescript fibrous tissue, or ligament, which underwent histopathological examination as macroscopically "suspected". The total number of such lesions recorded in a reported period of time was 15 (7.5% of the whole set).

Six specifically chosen, rare pathologies are presented below.

Table V. Metastatic tumours according to the source of dissemination

METASTATIC TUMOURS	NUMBER OF CASES
Lung	10
Breast	7
Kidney	2
Prostate	3
Pancreas*	1
Melanoma of the skin	1
Total	24

**Case of intramedullary metastatic carcinoma, pancreas being the most probable source (strong positivity for CK7 and CK20)*

Table VI. The frequency of the lesions' occurrence according to the region of the spine

REGION OF THE SPINE	NUMBER OF CASES	%
Cervical	23	12.43
Thoracic	116	62.71
Lumbar	46	24.86
Total	185	100

Case 1. Capillary haemangioma

Case presentation

A 79-year-old male with several years of pain history in the lower segment of the thoracic spine, numbness of the left leg and weakening of left leg muscles for the past two months. The neurological examination showed distal paresis of the left lower limb, loss of deep reflexes in lower extremities and the weakening of superficial sensation below the Th10 level on the left side. The MRI image showed the presence of a spinal canal tumour at the level of Th10-Th11. The laminectomy approach was performed on this patient during operation. The intradural tumour, located subarachnoideally on the back surface of the spinal cord, was removed. After treatment, transitional aggravation of left leg paresis was observed. During the long period control (after 6 months), the patient was walking unassisted but negligible left lower limb paresis remained. Pathologically, the lesion presented as multiple vascular channels lined with a single layer of endothelial cells strongly positive for CD34 (Fig. 1).

Short commentary

Capillary haemangioma is formed between the third and sixth week of foetal life whereas it becomes symptomatic most frequently in the fourth or fifth

decade. Its most common localization in the CNS is the pons [10]. In the spinal cord, it appears typically in the thoracic and lumbo-sacral region, most often intramedullary. It is rarely found on the surface of the spinal cord (like in our case in which the lesion was attached to the dura mater and adhered to the surface of the spinal cord). The basic indicated diagnostic method used is MRI. The capillary haemangioma might not be visible or barely visible in T1 and T2 sequences as a nidus of tubular structures, however, its signal enhances after contrast administration. The usual clinical manifestation, occurring commonly after physical effort, is spontaneous haemorrhage that presents without any previous neurological symptoms and leads to paresis or paralysis [8]. The histopathological differential diagnosis of capillary haemangioma should exclude reactive proliferation of endothelial cells and vessels.

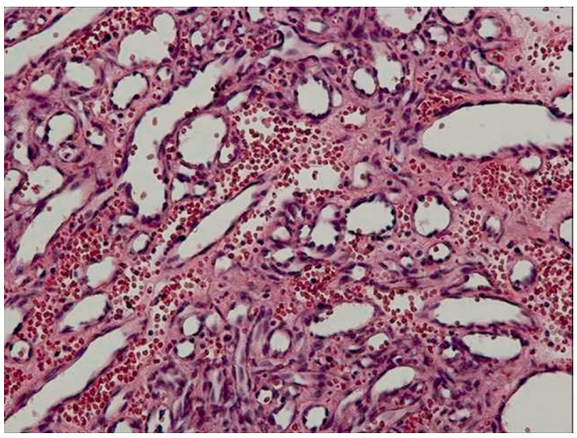

Fig. 1. Case 1, Capillary haemangioma. The tumour was broadly attached to the dura mater and adhered to the spinal cord. HE, magnification 200×

Case 2. Paraganglioma

Case presentation

For the past two months, a 64-year-old female suffered from lumbar pain radiating to the right lower limb and recently (two weeks) began radiating to both lower limbs. Seven years prior to admission, she had undergone an amputation of the right breast due to a carcinoma. Neurological examination revealed a positive Lasegue's sign by 50 degrees on the right side and a positive Mackiewicz sign on both sides. The MRI examination showed a lumbar spinal canal tumour (Fig. 2a). The patient was operated on by laminectomy at the level of L3. The tumour was localized intradurally on the nerve root and it was completely resected. Pathologically, the tumour presented with multiple, tightly compacted, uniform cells (in a histochemical examination, it was positive for chromogranin and synaptophysin) forming band-like structures and a trabecular pattern (Fig. 2b). There was no formation of 'Zellballen'. The tumour cells were NSE+ and GFAP–. Five days post operation, the patient was released from the hospital in good condition.

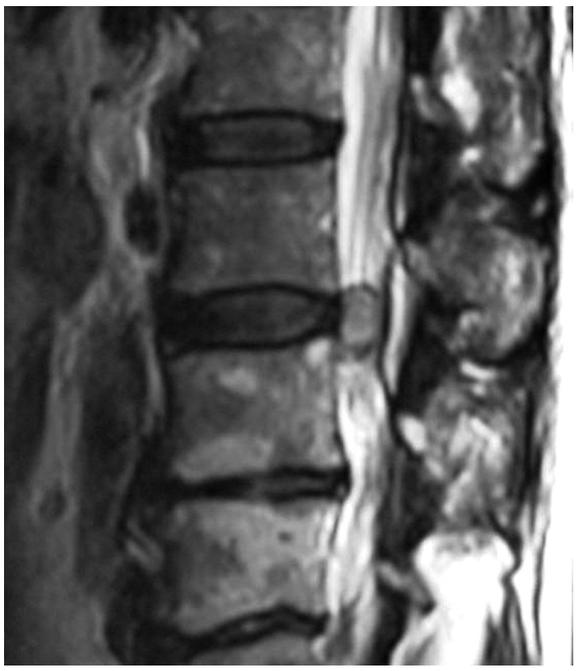

Fig. 2a. Case 2, MRI study of paraganglioma, T2 weighted image, sagittal view

Short commentary

Paraganglioma is a very rare tumour in the spinal canal. This kind of tumour is formed from paraganglia in the autonomic nervous system [11]. In the majority of cases, it occurs in the adrenal gland or glomus caroticum. Paraganglioma of the spinal canal is most frequently localized in the conus medullaris, cauda equina or filum terminale. The tumour is considered benign but may have various growth dynamics. Intratumoral haemorrhage is very frequent [8]. For a diagnosing neuropathologist, it is important to exclude the metastasis of a carcinoid. However, this is very difficult to establish and, in practice, it is most

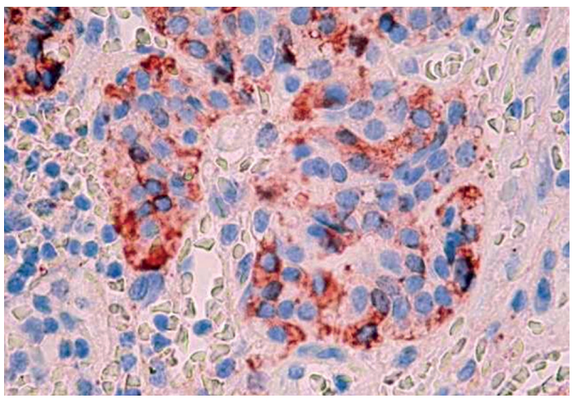

Fig. 2b. Case 2, Paraganglioma. Magnification 200×

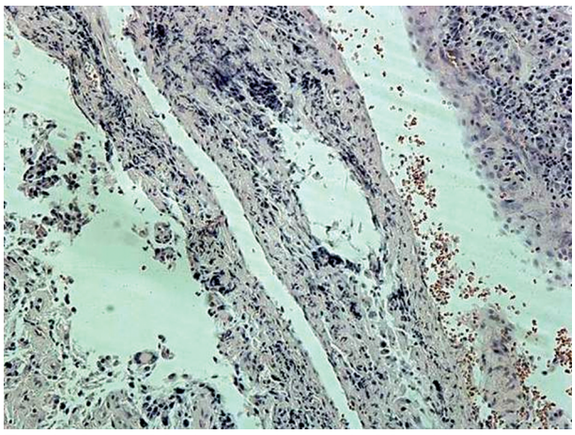

Fig. 3. Case 3, Meningeal gliomatosis. Magnification 100×

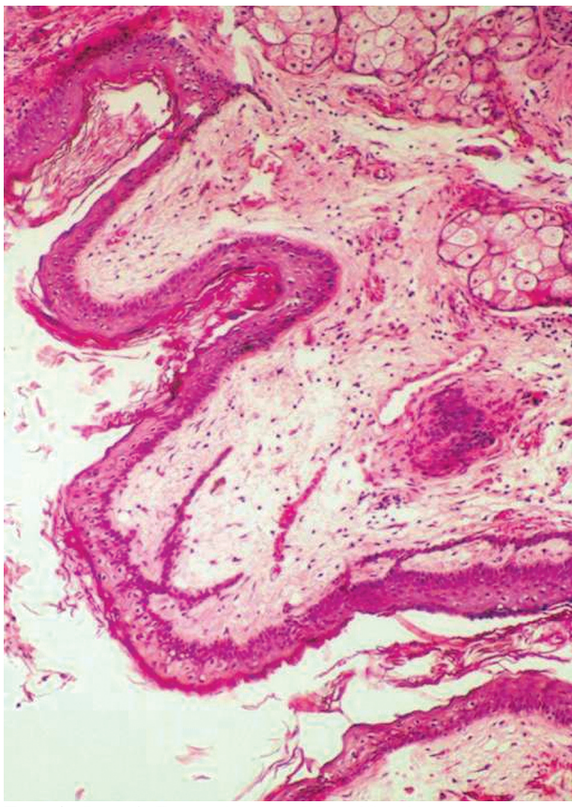

Fig. 4. Case 4, Dermoid cyst. Magnification 50×

important to initially check whether there are any symptoms that could indicate a carcinoid tumour.

Case 3. Meningeal gliomatosis

Case presentation

A 27-year-old patient suffered from sudden onset of headache, nausea and vomiting. Neurologically, he presented with neck stiffness. Lumbar puncture was performed and xanthochromic CSF emerged. A CT scan of the head did not show any features of haemorrhage. Upon admission to the neurosurgery clinic, the patient was conscious, had good verbal contact, neck stiffness, VI nerve paresis on the right side and hypoesthesia down from the Th11 level. The DSA (digital subtractive angiography) examination did not reveal any vascular malformation. An MRI study of the head and spine showed enhancement of the meninges with infiltration of the spinal cord, especially in the thoracic region. Partial resection of the intradural tumour, with spinal cord infiltration at the level of Th11, was performed. The histopathological examination showed a malignant GFAP positive glial tumour (WHO IV) (Fig. 3). Meningeal gliomatosis was diagnosed regarding the diffuse infiltration of the meninges in the spinal canal. Following the operation, lower limbs paresis appeared. The patient was qualified for chemotherapy.

Short commentary

Meningeal gliomatosis is a form of glioma associated with spreading to or multi-focal occupation of the meninges [12]. The disease occupies intracranial structures as well as the spinal canal. The prognosis is poor and the life expectancy, from the moment of diagnosis, is several weeks. Upon histopathological interpretation of the malignant glioma, which infiltrates the meninges of the spinal canal, one has to consider secondary dissemination of the intracranial or intramedullary glioma ("drop metastases") and "primary diffuse leptomeningeal gliomatosis" [13, 14]. The latter term is restricted to cases where there is no evidence of glioma in the brain or in spinal cord parenchyma. However, it is difficult to unequivocally prove the lack of existence of a parenchymal tumour.

Case 4. Dermoid cyst

Case presentation

A 42-year-old female patient complained of lower back pain radiating to both lower extremities, which persisted for the past two months. Neurologically, she presented positive Lasegue's sign by 40 degrees on both sides. The CT examination of the spine displayed a tumour located at the level of L1-L2. Surgery revealed a round yellowish tumour confined in the conus medullaris that compressed neighbouring roots. The tumour was partially resected and only its mass was reduced because it was not clearly separated from the neural tissue. A microscopic examination of the tumour revealed a polymorphic structure consisting of an agglomeration of epithelium, hairs, and lacunae of fat tissue. After the surgery, the patient presented no neurological deterioration. She was discharged in good condition. Histopathological assessment of the material showed characteristic features of a dermoid cyst (Fig. 4).

Short commentary

A dermoid cyst is a benign lesion that belongs to occult dysraphic disorders. It is created in the early stages of foetal life and it often affects children. It commonly coexists with the dermal sinus, which can lead to severe neurological complications caused by recurrent meningitis [15]. Dermoid cysts make up approximately 20% of intradural tumours in individuals in the first year of life. The dermoid cyst is most frequent localized intradurally in the lumbar region of the spine. Some of the dermoid cysts are acquired and may develop after the lumbar puncture or injury [16]. The tumour consists of skin elements like epidermis, fat tissue, keratin debris, hair, skin glands and their secretions. Calcifications may also occur. When the cyst ruptures, chemical meningitis may develop [17].

Case 5. Synovial cyst

Case presentation

A 79-year-old female suffered from lower back pain radiating to the right lower limb. For a one-month period, she had symptoms of neurogenic claudication. She was able to walk without pain for only a few meters. Neurologically, she presented with hypoesthesia around the right L5 nerve root dermatome, hypoactive knee jerk reflex on the right side, urinary incontinence, and negative stretch signs. The MRI examination showed features of lumbar degenerative stenosis. Laminectomy was performed at L4 and L5. The pathological mass was found attached to the facet joint at the level of L5 and it was completely removed. The symptoms were partly relieved after the operation. The pathological evaluation showed that the cystic cavity was covered with synovium positive for EMA (Fig. 5).

Short commentary

A synovial cyst is a benign lesion that develops as a result of degenerative changes in the intervertebral joints [18]. It is caused by the herniation of the synovial membrane outside of the joint capsule. It is commonly located at the level of L4/L5, in an area where there is the largest range of spinal movement [19, 20]. As the cyst increases in size, it may lead to the compression of nervous structures. The synovial cyst is formed by a thick collagen capsule, filled with a yellowish liquid, covered with villi on its inner side, and is connected to the intervertebral joint capsule [20].

Case 6. Intramedullary metastatic carcinoma

Case presentation

A 69-year-old woman suffered for the past two months from increasing weakening of the lower limbs, accompanied by urinary incontinence. Neurologically, she presented pyramidal paresis in the lower extremities, bilateral positive Babinski sign, and absence of the knee jerk and Achilles tendon reflexes. The MRI examination showed a T1 hyperintensive intramedullary mass, which was enhanced after

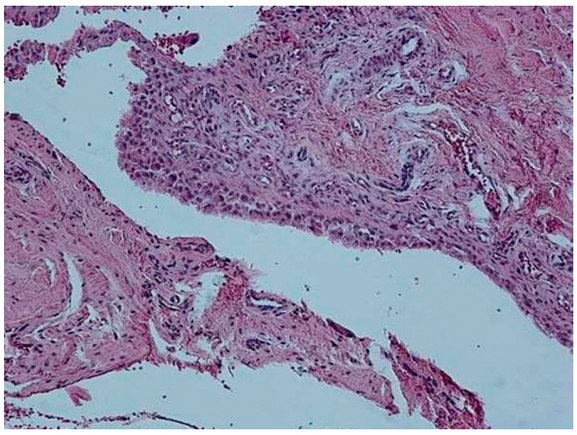

Fig. 5. Case 5, Synovial cyst. Magnification 50×

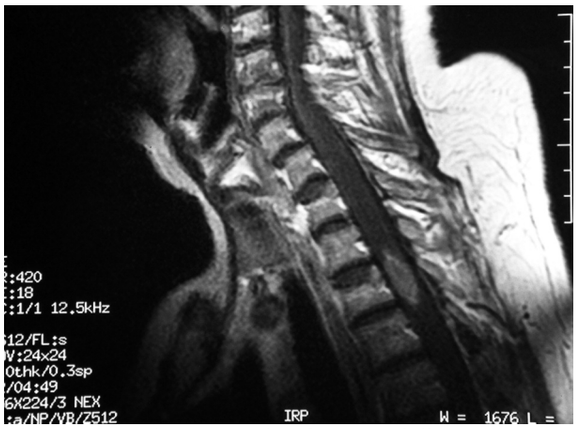

Fig. 6a. Case 6, MRI study of intramedullary metastatic carcinoma. T1 weighted image, sagittal view

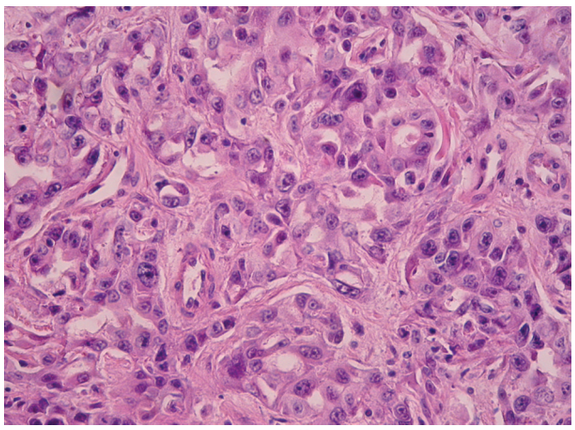

Fig. 6b. Case 6, Intramedullary metastatic carcinoma. Magnification 200×

contrast, at the level of Th2-Th4 (Fig. 6a). The tumour was completely removed. The patient's condition did not change following the surgery. The histopathological diagnosis of the mass was adenocarcinoma (Fig. 6b). The location of primary focus has not been definitely established, but the tumour turned out to be strongly positive for cytokeratins 7 and 20 and hence, though we cannot be sure, it seems that the most probable source of dissemination was pancreas.

Short commentary

In the case of carcinoma dissemination, the vertebral bodies are frequently affected [21], whereas intramedullary metastasis is an infrequent occurrence [22]. The intramedullary metastatic carcinoma constitutes about 2-5% of all metastatic tumours in the spine. The most common sources of intramedullary metastases are small cell carcinoma, breast carcinoma, melanoma and lymphoma. The symptoms develop earlier than in the case of extradural metastasis [5]. In our case, the source of metastasis is close to be defined as pancreatic or cholangiocarcinoma.

Discussion

The statistical cross-section of the types of spinal canal or spine tumours encountered depends on the character of the centre in which they were operated. In most neurosurgical departments, tumours of the spinal canal prevail. However, neurosurgeons and neuropathologists frequently face tumours of bone origin or neoplasms that do not originate from the spinal cord, meninges or nerves and which are not included in the WHO classification of the central nervous system tumours [1]. For this reason, it is difficult to find relevant statistical reviews on unclassified tumours. This paper reviewed material on the dominant spinal canal tumours but with some admixture of spine tumours to exemplify the spectrum of tumours that may be treated in a neurosurgical department. The material may not present a truly complete spectrum of tumours and other pathologies that can occur in this region. The statistics presented by other authors from orthopaedic centres are quite different. This can be exemplified by the work of Harms and Melcher, who also presented a ten-year study on spine tumours [23].

From a pathological point of view, rare lesions (like those presented above) pose a challenge for neuropathologists. This is shown especially in the case of meningeal gliomatosis, which emphasizes the necessity for a close cooperation of the neurosurgeon and the neuropathologist. In such cases, the diagnosis intrinsically contains both histopathological and clinical (neuroimaging) input.

The review of the material disclosed may in some cases be regarded as disputable. Some of the cases may contain errors made in preoperative diagnostics or surgical procedures resulting in histopathological findings like a spinal root, ligament tissue or fibrous tissue.

Our report does not include pathologies like nucleus pulposus or nonspecific findings like subdural hematoma, nonspecific inflammatory changes, or just samples of histologically apparently normal tissue like: fragments of spinal root, nondescript fibrous tissue, or ligament, which were subjected for histopathological examination as macroscopically "suspected". The total number of such lesions recorded in a reported period of time was 15.

Conclusion

The review shows the diversity of tumours of the spinal canal and spine and hopefully provides the practical insight into the spectrum of tumours in this anatomical region that could be encountered in a neurosurgical department. In such a setting, meningiomas prevail, followed by schwannomas and metastatic tumours.

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